Infrasellar Craniopharyngioma of The Posterior Nasal Septum: A Rare Entity

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SUMMARY

Objective: To report an unusual location of infrasellar craniopharyngioma in a peadiatric patient.

Case Report: A six-year-old boy presented with persistent bilateral nasal obstruction for one year. Clinical examination revealed a posterior choanal mass arising from septum and the finding was confirmed by paranasal sinuses computed tomography scan. He then underwent wide local excision. Histopathological examination confirmed the diagnosis of craniopharyngioma (adamantinomatous type). There were no signs and symptoms of recurrence after a year of follow-up.

Conclusion: Infracranial craniopharyngioma without sellar involvement is extremely rare. Persistent nasal obstruction without endocrine dysfunction is the common presentation. Radiological imaging is important to diagnose and assess the extent. The mainstay of treatment for infrasellar craniopharyngioma is surgery. Regular follow up is mandatory.

KEY WORDS:
Craniopharyngioma, Infrasellar, Septum

INTRODUCTION

Craniopharyngioma is a benign intracranial tumour. It is rare but not uncommon. The incidence is 0.13 cases per 100,000 person-years. It accounts for 2-5% of all the intracranial tumours in overall and 5.6-15% of all the tumours in paediatric groups. There is a bimodal age distribution with the first peak at five to 14 years and a second peak at 50 to 74 years [1]. Despite of its histological benign features, it can behave aggressive locally and associated with frequent recurrence. Craniopharyngioma is typically located within sellar and suprasellar region. Craniopharyngioma with infrasellar extension is rare whereas craniopharyngioma without sellar involvement is extremely rare entity. Herein, we report a case of craniopharyngioma presenting as a mass at posterior nasal septum in a six-year-old boy.

CASE REPORT

A six-year-old boy presented to otorhinolaryngology clinic, complaining of persistent bilateral nasal obstruction for one year without any other nasal and ear symptoms. Clinical examination revealed a posterior choanal mass arising from septum. Other ear, nose and throat examinations revealed insignificant findings.

Computed tomography of paranasal sinuses showed heterogeneously enhancing mass arising from posterior nasal septum measuring 1.8x0.8cm. (Fig.1) There was no bony erosion or extension into paranasal sinuses or intracranial.

He then underwent examination under anaesthesia. The examination showed an exophytic nasal mass arising from the posterior part of the septum involving choana while the nasopharynx was clear. Wide local excision and biopsy was performed at the same setting.

Histopathological examination revealed a solid neoplasm composed of proliferating odontogenic like epithelium with loosely cohesive central stellate reticulum-like cells and peripheral palisaded cuboidal or columnar pre-ameloblast-like cells. Juxtaposed between the tumour epithelium are irregular masses of ghost cells. Stromal cysts and foci of calcifications are observed. (Fig.2) The surgical margin was clear. The diagnosis of craniopharyngioma (adamantinomatous type) was made.

Post-operatively, the child was well and the nasal obstruction was relieved immediately. He is still on regular follow up in otorhinolaryngology clinic without any symptoms of recurrence.

DISCUSSION

Craniopharyngioma is a rare intracranial tumour. The tumour arises from remnants of the craniopharyngeal duct or Rathke cleft. It is typically located at sellar or suprasellar region. Infrasellar extension is rare as the expansion is limited by sphenoid bone that acts as a boundary. There are approximately 40 reported cases of craniopharyngiomas with infrasellar extension.2 Infrasellar craniopharyngioma without sellar involvement is extremely rare. There are only six cases reported in literature. All the studies are conducted in case study format. The sites reported are sphenoid sinus, ethmoid sinus, maxillary sinus and nasopharynx [2, 3]. Unlike suprasellar craniopharyngiomas, nasal blockage is the prominent feature in infrasellar craniopharyngiomas without sellar involvement[4]. All the reported cases show no endocrine dysfunction [3]. Our case is one of the infrasellar

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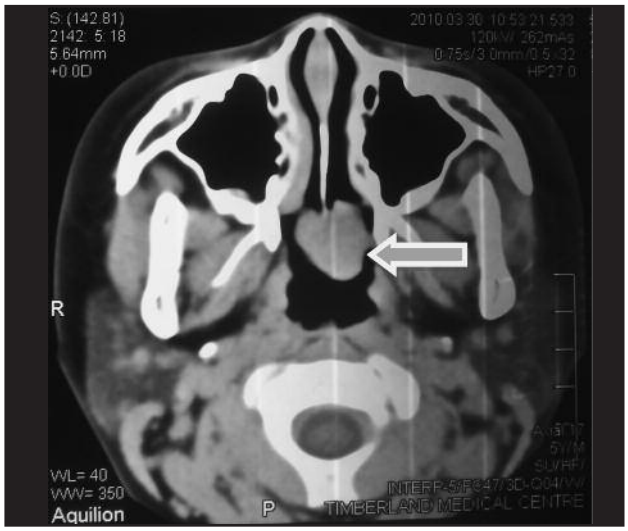

Fig. 1: Computed tomography of paranasal sinuses showed heterogeneously enhancing mass arising from posterior nasal septum measuring 1.8x0.8cm.

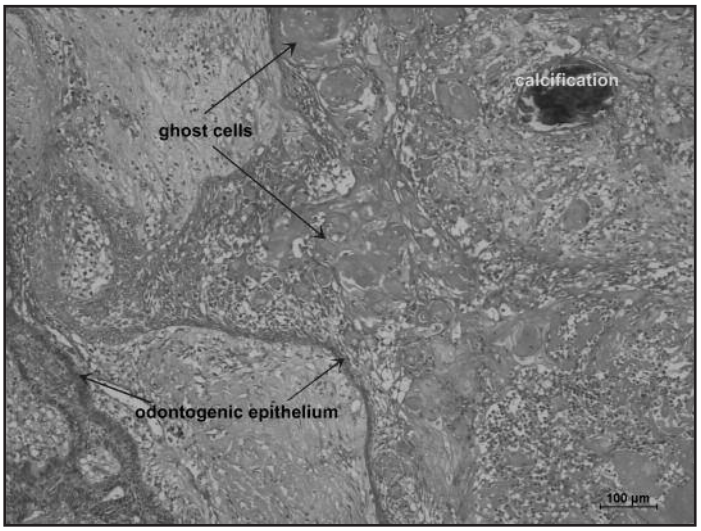

Fig. 2: A solid neoplasm composed of proliferating odontogenic like epithelium with loosely cohesive central stellate reticulum-like cells and peripheral palisaded cuboidal or columnar pre-ameloblast-like cells.

craniopharyngiomas without sellar involvement. The unusual location at posterior septum has not been reported so far. As like other infrasellar craniopharyngiomas, the child presented with nasal blockage without endocrine dysfunction.

The diagnosis of craniopharyngioma is based on tissue biopsy. Histologically, craniopharyngioma is categorized into adamantinous and squamous-papillary form. The former is typically found in paediatric group whereas the latter normally found in adult. In adamantinomatous craniopharyngioma, histology will show a typical adamantinomatous epithelium in palisading pattern, surrounded with stellate cells, wet keratin, keratohyaline granules and other components like cysts, cholesterol, inflammation, giant cell reaction and calcification [5]. Ameloblastoma is one of the important differential diagnosis for infrasellar craniopharyngioma without sellar involvement. Ameloblastoma is a benign odontogenic tumour usually found in mandible. Craniopharyngiomas and ameloblastoma share identical histological features and the only way to differentiate these two conditions are based on its anatomical location. 3 We encounter difficulty to establish the diagnosis in this case. The presence of ghost cells favours the diagnosis of dentinogenic ghost cell tumour, which is a variant of ameloblastic tumour. However, in view of the location of the tumour, the diagnosis of craniopharyngioma is made after discussion with pathologists.

Computed tomography scan (CT) and magnetic resonance imaging(MRI) are two useful radiological tools to delineate the tumour and formulate the management. 2 Craniopharyngioma shows cystic and solid component in both imaging. CT scan can depict the bony anatomy and calcification clearly. MRI is particularly beneficial for intracranial craniopharyngioma. MRI is valuable in assessing the structure and features of the tumour in relation to the surrounding soft tissue. However, MRI is not as good as CT to detect calcification. 1 In our patient, CT scan was done and showed benign features that were consistent with craniopharyngioma.

The mainstay of treatment for craniopharyngioma is surgery with or without radiotherapy. The management of intracranial craniopharyngioma is always controversial due to its anatomical location near to pituitary gland. However, for infrasellar craniopharyngioma, complete excision is more feasible. The approach can be done transpalatal, transnasal, or lateral rhinotomy depending on the location of the tumour. Complete excision without radiotherapy is the main aim. Complete excision reduces the recurrent rate and avoids the complication of radiotherapy. 2 In fact, the extent of the surgical resection is the single most significant factor associated with recurrence. In our patient, the decision of complete excision was decided as the appearance of tumour raised the suspicion of malignancy intraoperatively. Furthermore, the tumour clear margin was confirmed by histopathological examination. He remained symptom free after a year of follow up.

CONCLUSION

Infracranial craniopharyngioma without sellar involvement is extremely rare. Persistent nasal obstruction without endocrine dysfunction is the common presentation. To date, there is no reported infrasellar craniopharyngioma case at posterior nasal septum. Radiological imaging is important to diagnose and assess the extent. The mainstay of treatment for infrasellar craniopharyngiomas is surgery. Regular follow up is mandatory.

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